

# Achalasia Carcinoma Sequence

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## ABSTRACT

*We report a case of carcinoma of the esophagus in a 58 years old woman with achalasia, who has been diagnosed since 30 years ago, which initiated by surgical treatment (myotomy) and the symptoms recurred since 3 years ago.*

*According to the progress of the disease, malignancy was strongly suspected due to prolonged stasis and mucosal irritation caused by achalasia (achalasia carcinoma sequence). Because of these contributing factors for the development of serious complications such as malignancy, the diagnosis of achalasia must be systematically diagnosed and treated aggressively. Surveillance endoscopy in patients with achalasia should be performed every 1-2 years.*

**Key words:** achalasia carcinoma sequence, surveillance endoscopy

## INTRODUCTION

Achalasia is a disease in which there is a double defect in esophageal function, including inappropriate lower esophageal sphincter (LES) relaxation and loss of peristalsis in the lower two thirds of the smooth muscle portion of the esophagus.

We can detect abnormalities in muscle and nerve component:

- Loss of ganglion cells within the myenteric (Auerbach) plexus (the most important).
- Degeneration of the vagus nerve.
- Qualitative and quantitative changes in the dorsal motor nucleus of the vagus.

The first case was reported by Sir Thomas Willis in 1672, and the term of achalasia was coined by Hurst and Rake in 1929 who introduced bougie dilator in the treatment of achalasia.

Between 1998-2000, there were 26 new cases of achalasia who came to the Division of Gastroenterology, Department of Internal Medicine, Cipto Mangunkusumo National General Hospital Jakarta, two of them were twins.

All of the patients had solid food dysphagia and most of them had variable degree of liquid food dysphagia. Other complaint were regurgitation, chest pain, heart-burn and weight loss.

Diagnosis was made by physical examination (although rare abnormalities were found), radiography

(barium meal) and endoscopy (with biopsy). Endoscopic examination is always required to exclude a neoplastic process at the location of the gastroesophageal junction, and to evaluate the esophageal mucosa before therapeutic manipulation. The diagnosis of achalasia, if available, should be established by esophageal manometry.

Treatment:

- diet
- pharmacotherapy (anticholinergic, isosorbide dinitrate, calcium channel blocker)
- esophageal dilatation (Hurst bougie, Savary bougie, pneumatic/ballon dilator)

If conservative treatment is failed, surgical treatment (myotomy) should be considered.

The newer conservative treatment:

- Transcutaneous electric nerve stimulation (TENS)
- Injection of Botulinum toxin

## CASE REPORT

A Sundanese 58 years old woman presented with the complaint of dysphagia, especially solid food, since 3 years ago.

Actually, she has been diagnosed as achalasia since 1970 and at that time surgery (myotomy) was performed. After the surgery, the complaints were completely relieved. Since 3 years ago, she has experienced the same complaints and it much worsened in the last year with the loss of body weight of 20 kg.

Physical examination: malnourished (body weight was 28 kg, height 150 cm) and conjunctiva were pale. No other abnormalities were found.

Haemoglobin level was 4,8 g/L. Barium meal examination revealed a very dilated middle and lower part of esophagus (sigmoid type).

Endoscopy: LES was very stenotic with dilatation of esophageal lumen from the middle until the lower part. The mucosa was fragile and nodular. Biopsy was performed and histopathological findings revealed squamous cell carcinoma.

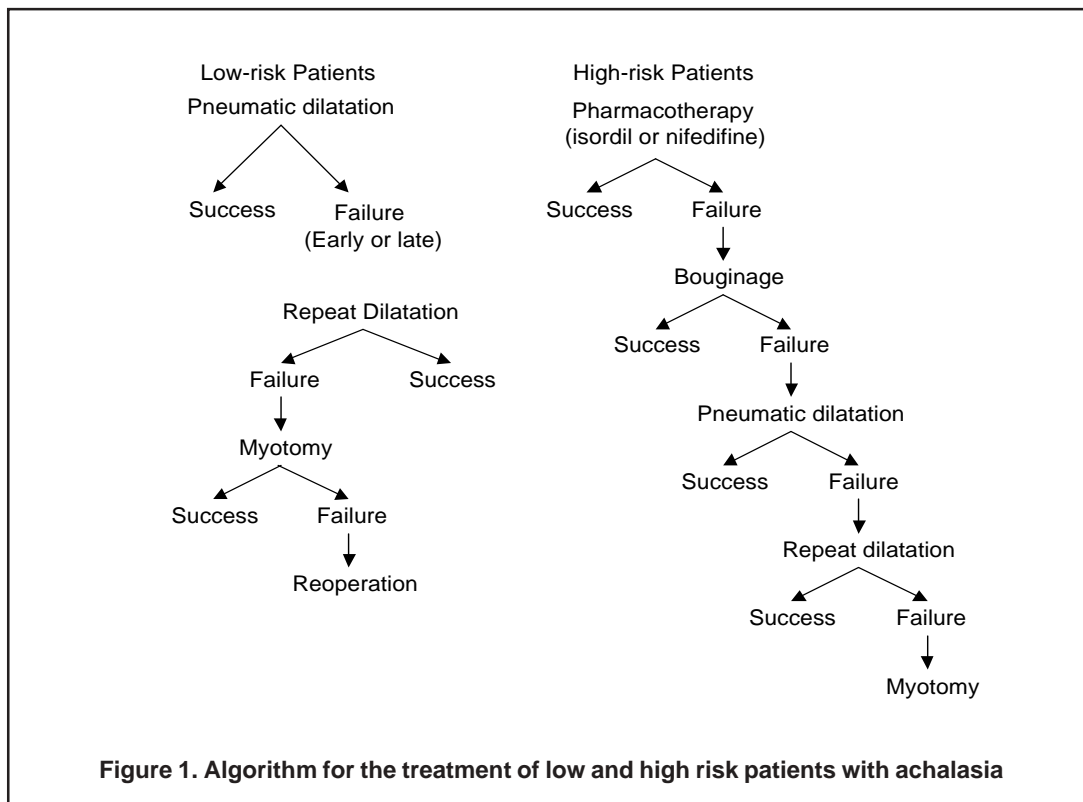
Dilatation by balloon/pneumatic dilator was performed 3 times, that resulted in a relieve of symptoms. The patient refused surgical treatment.

## DISCUSSION

Achalasia is uncommon but not rare. In this patient, achalasia was already diagnosed 30 years ago, in which initiated by surgical treatment. The symptoms started to

return 3 years ago, and worsened in the last year. The last investigation revealed a marked esophageal dilatation and the clear appearance of malignancy (squamous cell carcinoma). Because of the progress of the disease, the development of carcinoma in this patient was strongly suspected due to prolonged stasis and mucosal irritation caused by achalasia (achalasia carcinoma sequence). The anemia occurred due to deficiency. Because marked esophageal dilatation and stasis may be important contributing factors for the development of serious complications, the diagnosis of achalasia must be systematically diagnosed and aggressively treated. Surveillance endoscopy in patients with achalasia should be performed every 1-2 years.

Algorithm for the treatment of low and high risk patients with achalasia as follows.



## CONCLUSION

We report a case of carcinoma of the esophagus in a patient with achalasia, who has been diagnosed 30 years ago, which initiated by surgical treatment (myotomy).

The symptoms recurred 3 years ago and the investigation revealed a marked dilatation of the esophagus and stenosis of LES and a clear appearance of esophageal malignancy.

According to the progress of disease, this malignancy was strongly suspected due to prolonged stasis and mucosal irritation caused by achalasia (achalasia carcinoma sequence).

## REFERENCES

1. Hewson EG, Ott DJ, Dalton CB, et al. Manometry and radiology complementary studies in the assessment of esophageal motility disorder. *Gastroenterology* 1990; 93: 3.
2. Kahrilas PJ, Kishle SM, Helm JF, et al. Comparison of pseudoachalasia and achalasia. *Am J Med* 1987; 82: 439.
3. Howard PJ, Maher L, Pryde A, et al. Five years prospective study of the incidence, chemical features and diagnosis of achalasia in Edinburgh. *Gut* 1992; 33: 1011.
4. Coccia G, Bortolotti M, Michetto P, Dedoro M. Prospective clinical and manometric study comparing pneumatic dilatations and sublingual nifedipine in the treatment of esophageal achalasia. *Gut* 1991; 32: 604.
5. Eckardt VF, Aigner C, Bernhard G. Predictors of outcome in patients with achalasia treated by pneumatic dilation. *Gastroenterology* 1992; 103: 1732.
6. Richter JE. Motility disorders of the esophagus in Yamada T, et al (eds); *Textbook of Gastroenterology*, JB. Lippincott Company Philadelphia 2<sup>nd</sup> edition 1995: 1174-1212.